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KIMURA'S DISEASE-A RARE CAUSE OF LYMPHADENOPATHY

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ABSTRACT

Kimura's disease is a rare inflammatory disorder of unknown cause, primarily seen in young Asian males. The disease is characterized by a triad of painless subcutaneous masses in head and neck region, blood and tissue eosinophilia and moderately elevated serum IgE levels. Here, we present a case report of this rare entity with classical histopathogical features.

KEYWORDS: FNAC, Pyogenic Granuloma, CD4, Epithelioid Haemangioma, ALHE

INTRODCUTION

Kimura's disease is a rare entity causing subcutaneous swellings and lymphadenopathy, with hardly 120 cases reported worldwide. [1] It is mainly seen in Asian countries. We are reporting the case of a 7-year old child affected by this rare disease. Even though it is a benign disease in a majority of cases, it can produce devastating renal and thrombotic complications [2] Therefore, it needs proper follow-up.

Case Report

A seven year old boy presented with swelling behind left ear for 2 years. It was painless, insidious in onset, gradually progressive, localized to the left post auricular region. Subsequently he developed a similar swelling behind right ear after 3 months. There was no other associated symptoms. No contact h/o of TB documented. Past history and family history were uneventful. Examination showed a solitary swelling situated in the occipital area left side 2x2cm, oval in shape ,well defined edges with no local rise of temperature or tenderness, smooth surface, borders have well defined, firm in consistency, plane of swelling-subcutaneous suggestive of lymph node. Other lymph nodes is not enlarged. Other system examination was

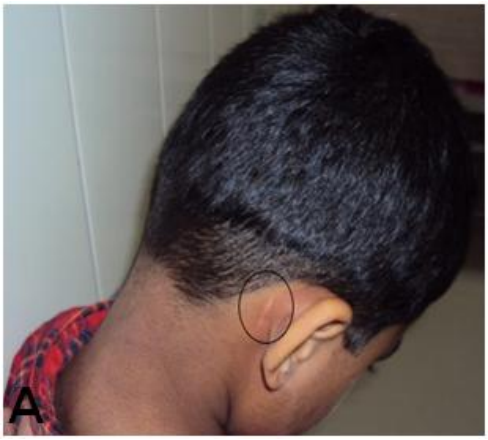

Figure A: Picture Showing Excised Scar of Lymph Node in Preauricular Region

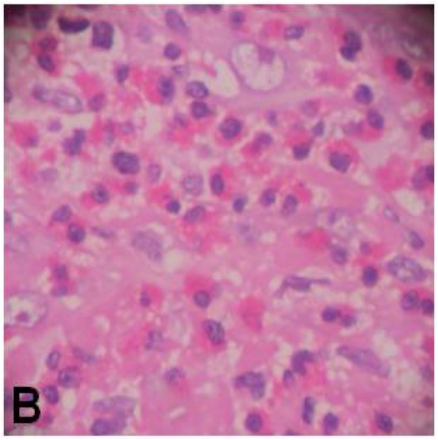

Figure B: Section of Lymph Node Showing Dense Eosinophilic Matrix, Giant Cells, Vascular Proliferation

Investigations showed that, elevated counts with eosinophilia. TB workup was negative. In view of reactive adenitis suggested by FNAC, he was received treatment for infective etiology with antibiotics. Since patient was not response, an excision biopsy was done. Histopathology examination showed features of lymph node with dense eosinophils, scattered plasma cells, giant cells, granulomas, reed sternberg like cells and vascular proliferation - features are in favour of kimura's disease. Patient had received oral steroids for 4 weeks following which size of the swelling regressed.

DISCUSSIONS

Kimura's disease was first reported by Chinese authors (Kimm and Szeto in 1937) [1] and has been variously known as epithelioid haemangioma, atypical pyogenic granuloma and cutaneous eosinophilic lymphofolliculosis. The defined characteristic featureswas published by Kimura's et al.,in Japan in 1948.[2] There had been some 120 cases were described in the world literature. The majority of patients have originated in the Eastern hemisphere particularly Asians.[1]

Its pathogenesis involves T-cell immunoregulation induced IgE-mediated type 1 hypersensitivity resulting in the release of eosinophiliotrophic cytokines secondary to allergy, infection or trauma.[3] The Persistent antigenic stimulus causes marked proliferation of human leukocyte antigen-DR. CD4 cells resulting in abnormal proliferation of lymphoid follicles. Activated CD4 cells of the TH-2 phenotype thus release cytokines such as granulocyte macrophage colony-stimulating factor and tumor necrosis factor, interleukins (IL-4, IL-5, IL-13).[4] This in turn may precipitate the high serum IgE and marked eosinophilia.

Pathology is characterized by prominent germinal centers in involved lymphnodes containing cellular, vascular, and fibrous components. The cellular component consists of dense eosinophilic infiltrates in a background of abundant lymphocytes and plasma cells, eosinophilic microabscesses with central necrosis, Warthin-finkeldey-type polykaryocytes, some degree of vascular proliferation of germinal centers.[5]Clinical features are characterized by a triad of Painless unilateral cervical adenopathy or subcutaneous masses predominantly in the head or neck region, Blood and tissue eosinophilia andMarkedly elevated serum immunoglobulin E (IgE) levels.[6]

The clinical course of kimura's disease is generally benign and self-limited. Usual presentation is typically slow-growing, painless masses with occasional pruritus. It involves subcutaneous tissues, lymph nodes (periauricular, axillary, and inguinal) parotid and submandibular glands.[7] Other unusual sites of involvement include the auricle,scalp, and orbit and has no malignant potential. Its main concern is disfigurement also known to recur following medical or surgical treatment. The lesions of kimura's disease usually preceed or coincide with the development of renal disease such as nephrotic syndrome or nephritic picture.[8]

Complete Blood Count almost always reveals peripheral eosinophilia (98%).[6] Serum immunoglobulin E levels are often elevated. Surgical biopsy is the most frequent diagnostic procedure. Kimura's disease can mimic other disorders such as Tuberculous lymphadenopathy, Angiolymphoid hyperplasia with eosinophilia (ALHE), Mikulicz's disease, eosinophilic granuloma, Malignant lymphoma, Salivary gland tumors.[9] Disease can be often confused with tubercular lymphadenopathy in countries where tuberculosis is most common but the typical feature of marked eosinophilia in histopathology examination with eosinophilic micro-abscesses and polykaryocytes help to differentiate from TB lymphadenopathy.

The event ofdisease may also be confused with ALHE but it typically presents in women during early to mid-adult life, lymphadenopathy is uncommon, eosinophilia is noted in <10% of cases. Histologically, the presence of inflammation

around medium-sized arteries or veins, evidence of vascular damage [florid fibrointimal proliferation], cuboidal to dome-shaped endothelial cells are seen.[9]

The Rx involves intralesional or oral steroids.[6] Radiotherapy has occasionally has been used to treat recurrent or persistent lesions. Cyclosporineis being reported to induce remission in patients with kimura's disease.[10] Other agents is tried are Oral pentoxifylline, All trans -retinoic acid in combination with prednisone, Imatinib.[10] Surgical biopsy should cause regrowth which can be managed medically.

The prognosis for kimura's disease is good with no potential for malignant transformation. Course ofdisease is a chronic and lesions frequently persisting or recurring despite treatment.[10]

CONCLUSIONS

In developing countries where, TB is a commonest manifestingas a chronic lymphadenopathy in head and neck region .Especially in children, Kimura's disease can be considered in cases of cervical lymphadenopathy with negative TB workup.

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